Pediatrics and Neonatology (2016) xx, 1–2

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BRIEF COMMUNICATION

Intramuscular Venous Malformation in an Infant Masquerading as Recurrent Gonarthritis

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Received Aug 11, 2015; received in revised form Oct 6, 2015; accepted Oct 30, 2015
Available online ■ ■ ■

1. Introduction

Vascular malformations, commonly misdiagnosed and mistreated as hemangiomas, are rare congenital abnormalities in the vasculature. Venous malformation (VM) is one of the most common vascular malformations and is easily diagnosed by skin discoloration when it involves the skin or superficial tissue. However, deep-sited VM may only manifest as a subcutaneous mass or can even be asymptomatic.[1,2] Particularly in patients with predominantly intramuscular VM, symptoms often develop later in life as physical activity induced pain with no fever or inflammatory responses emerging except in the case of infectious complications. Herein, we present a rare infantile case of intramuscular VM mimicking gonarthritis.

2. Case Report

A 10-month-old female infant was referred to us because of recurrent monoarthritis. The patient was first hospitalized at age 5 months with fever and swelling in her left thigh and knee. Leukocyte count was 23.4 $\times$ 10^9/L. Diagnosed with gonarthritis, the patient underwent antibiotic therapy and soon her condition improved. Thereafter, she experienced recurrent joint swelling accompanied by fever at ages 6 months, 7 months, and 8 months. Despite high leukocyte counts (16.0–27.4 $\times$ 10^9/L) and elevated levels of C-reactive protein (2.8–15.0 mg/dL) at these recurrences, no causative bacteria were detected. This infant was transferred to our tertiary hospital upon the fifth recurrence.

On admission, the patient's body temperature was 37.0°C. Physical examination revealed a fretful and anemic infant. The lower half of the left thigh and knee joint showed several symptoms of inflammation including calor with restricted movement. Complete blood counts yielded a leukocyte count of 20.2 $\times$ 10^9/L with 52% neutrophils and 37% lymphocytes, a hemoglobin concentration of 7.0 g/dL, and a platelet count of 553 $\times$ 10^9/L. In addition to normal creatinine kinase levels, blood tests revealed normal liver and kidney functions. C-reactive protein level was 3.98 mg/dL. Serum procalcitonin level was $<$ 0.1 ng/mL. Coagulation studies were unremarkable except for slightly increased D-dimer levels (2.6 μg/mL; reference range, 0–1.0). Cultures of peripheral blood and synovial fluid were negative for bacteria. Without antibiotics, the patient spontaneously became convalescent. Following remission, magnetic resonance imaging (MRI) revealed a well-circumscribed, partially lobulated mass in the vastus lateralis with high signal intensity on T2-weighted images (Figure 1A) and low signal intensity on gadolinium enhanced T1-weighted images (Figure 1B). Dynamic time-resolved magnetic resonance (MR) angiography showed diffuse enhancement of venous channels with hampered blood flow in the same site (Figure 1C–E).

Surgical intervention revealed a multilobular tumor located in the vastus lateralis surrounded by fibrotic tissue containing old thromboses. The lesion was completely removed. Histopathological analysis showed dilated postcapillary venules and veins between skeletal muscle bundles (Figure 1F). Massive sclerotic changes associated with accumulation of hemosiderin-laden macrophages were present. These findings led to the diagnosis of intramuscular VM. The patient did not experience a recurrence within 2 years of surgical resection.

3. Discussion

A female infant patient was presented with five recurrent febrile bouts mimicking septic monoarthritis of the knee. Histopathological analysis

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http://dx.doi.org/10.1016/j.pedneo.2015.10.013

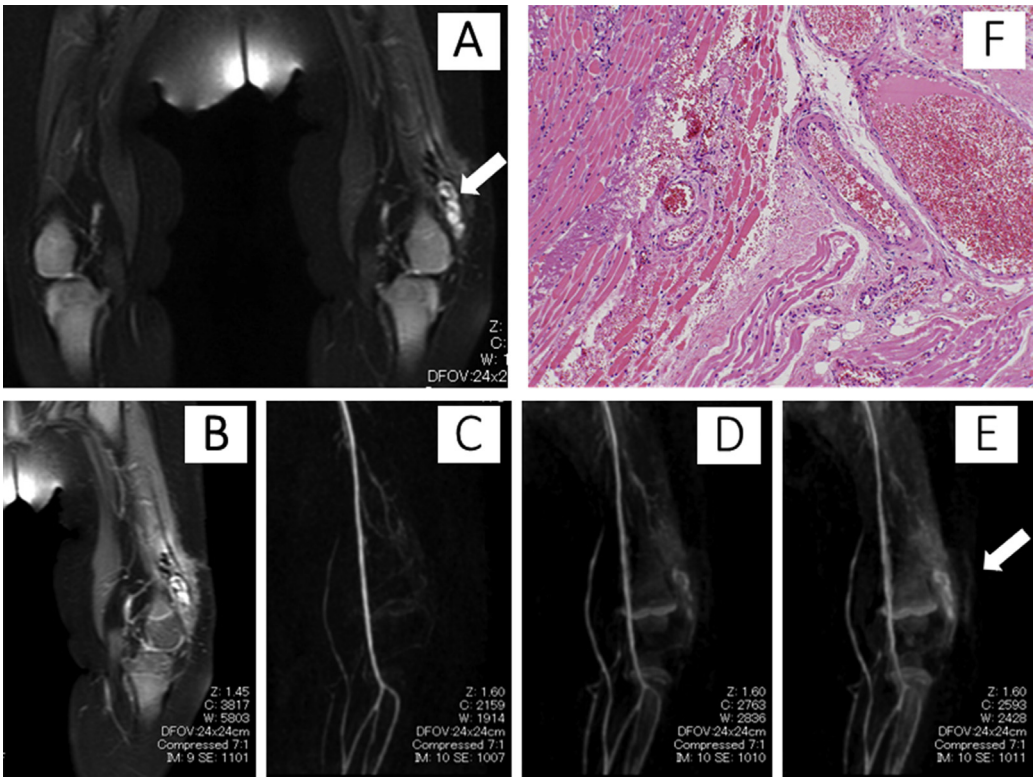

Figure 1 (A) Fat-suppressed T2-weighted image and (B) gadolinium-enhanced fat-suppressed T1- weighted image. A well-circumscribed, partially lobulated mass in the vastus lateralis with a maximal diameter of 2.4 cm (*arrow*). (C—E) Dynamic time-resolved MR angiography showed a paucity of arterial flow-related enhancement (50 seconds, 80 seconds, and 120 seconds after infusion of contrast medium, respectively). (F) Histopathological findings revealed dilated postcapillary venules and veins between skeletal muscle bundles (hematoxylin and eosin stain at 100× magnification), accompanied by hemosiderin-laden macrophages and massive sclerotic changes (not shown).

of the surgically resected mass in the left thigh showed an accumulation of siderophages, indicating frequent bleeding and associated inflammation. Intramuscular VMs are often associated with a history of trauma, and cause pain and swelling that are exacerbated by physical activity.[1,3] Considering the presence of a sclerotic lesion in the left thigh of the anemic infant, this unique clinical presentation may be explained by intramuscular hemorrhaging due to tissue injury. Excessive loads on the muscles and joints of the lower limbs may have precipitated these injuries when the patient began to crawl during infancy. Recurrent bleeding leads to local and systemic inflammation secondary to the extravascular accumulation of heme and heme-derived iron that are pro-oxidant, proinflammatory, and can be directly cytotoxic.[4]

Intramuscular VM is often misdiagnosed as intramuscular hemangioma. Since effective treatment depends on accurate diagnosis, improper diagnosis may lead to ineffective treatment. In brief, hemangiomas are benign vascular tumors that develop in three phases: a proliferative phase characterized by rapidly dividing endothelial cells, an involuting phase, and an involuted phase characterized by complete regression.[3,5] In comparison, VMs arise from dysplastic venous channels that gradually infiltrate normal tissue and progress without involution.[3,5]

The diagnosis and treatment of intramuscular VM in infancy is challenging because of its rarity and wide range of clinical presentations. MRI in combination with MR angiography is useful for diagnosing and evaluating the extent of vascular lesions. Following arthrocentesis, MRI at the time of the initial febrile bout revealed a mass in the vastus lateralis, indicating intramuscular hemorrhage. Nevertheless, accumulation of excess synovial fluid, systemic inflammation, and previous interventions made it difficult to distinguish septic arthritis, idiopathic monoarthritis, and periarticular intramuscular bleeding from VM. Furthermore, active bleeding around the intramuscular VM during the acute bleeding phase masked the hemorrhagic origin in previously acquired MR images. In summary, MRI is strongly recommended during the nonactive bleeding phase for the accurate diagnosis of VM.

Conflicts of interest

The authors have no conflicts of interest relevant to this article.

Acknowledgments

We thank Dr Takeshi Kamitani (Department of Clinical Radiology, Graduate School of Medical Sciences, Kyushu University, Fukuoka, Japan) for giving advice on MRI.

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Please cite this article in press as: Nanishi E, et al., Intramuscular Venous Malformation in an Infant Masquerading as Recurrent Gonarthritis, Pediatrics and Neonatology (2016), http://dx.doi.org/10.1016/j.pedneo.2015.10.013